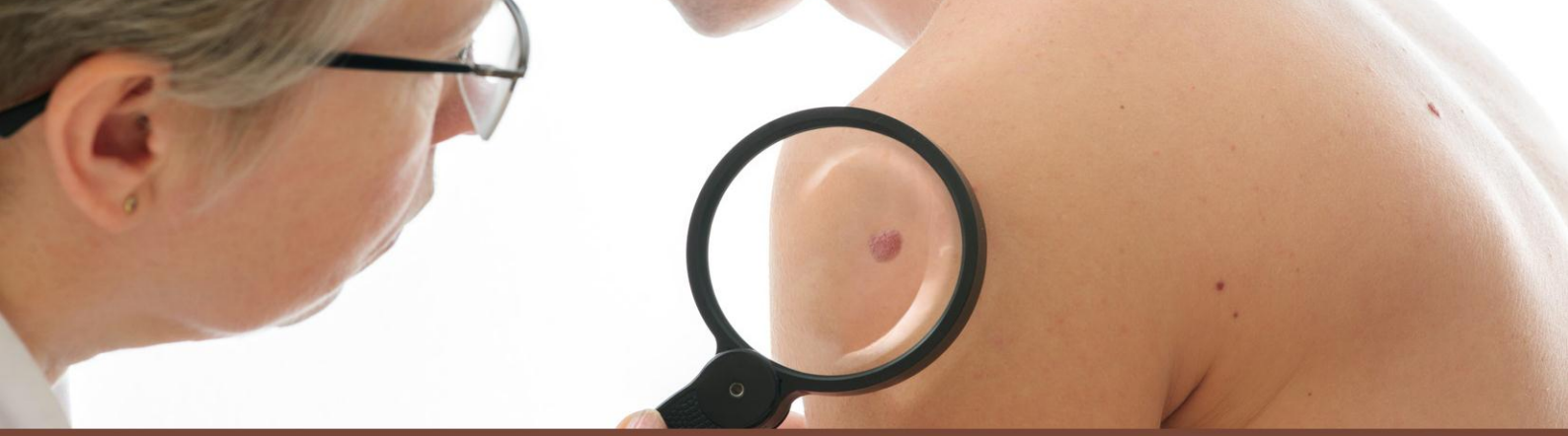

433 Teams

DERMATOLOGY

Lecture (6)

Other Connective tissue Disease

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Objectives:

- To learn how to diagnose and investigate dermatomyositis.
- How to manage dermatomyositis.
- To learn the presentation of morphea and systemic sclerosis and
- ways to manage them.
- To recognize other diseases like rheumatoid nodules and mixed CTD.
- This lecture is not meant to be inclusive of all the information about these diseases but to highlight important aspects in their diagnosis and management.

Color index: **slides,** **doctor notes,** **432 notes**

Dermatomyositis

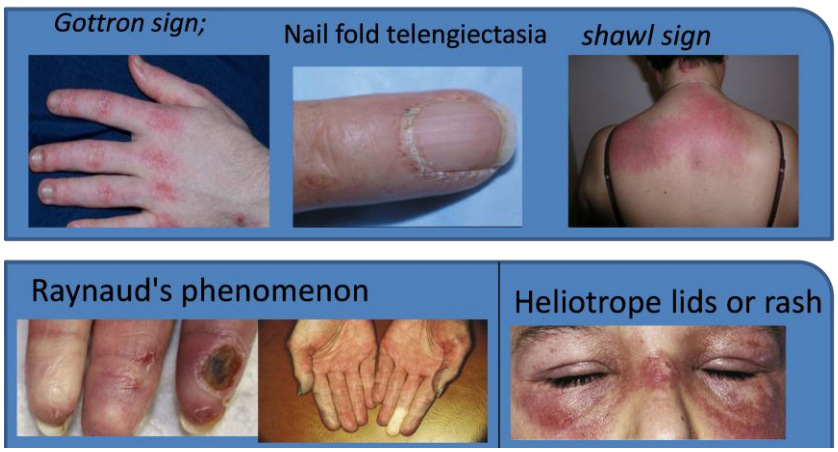
- Uncommon group of diseases with loss of muscle strength secondary to autoimmune muscle damage.
- affects adults between 40-60 (females mainly) and children 5-15.

- **Skin changes**: A violet-colored or dusky red rash on face and eyelids and on areas around nails, knuckles, elbows, knees, chest and back. The rash, which can be patchy with bluish purple discolorations, is often the first sign of dermatomyositis. periorbital & eyelid edema (**heliotrope lids**), lichenoid papules over finger joints & knuckles (***gottron sign***), erythematous macules & plaques on upper back, shoulders & back of neck (*shawl sign*)
**V-sign* similar to shawl but appears on the front of chest.

- Heliotrope rash: a violaceous -to-dusky erythematous rash with or without edema in a symmetrical distribution involving periorbital skin.
- Muscle weakness: Progressive proximal muscle weakness involves the hips, thighs,shoulders, upper arms and neck. The weakness is symmetrical and more in the extensor muscles. (in progressive cases patients use wheelchair)

Other signs and symptoms include:

- Photosensitivity
- Raynaud's phenomenon
- Nail fold telengiectasia
- Dysphagia, gastrointestinal ulcers
- Muscle pain or tenderness



It can be associated with: Other connective tissue diseases such as lupus, rheumatoid arthritis, Scleroderma and Sjogren's syndrome.

Cancer, Especially in older patients, particularly of the cervix, lungs, pancreas, breasts, ovaries and gastrointestinal tract. Cancer could precede, coincide or follow the diagnosis of DM.

Classification:

1- *Polymyositis*

2- *Adult dermatomyositis*: typical onset 40 yrs.

3- *Juvenile dermatomyositis*: more acute with vasculitis (GI hemorrhage) later calcification, lipodystrophy.

4- *Paraneoplastic dermatomyositis*: later onset associated with breast, head & neck carcinomas.

5- *Amyopathic dermatomyositis*: DM with no evidence of muscle involvement over 6 months follow up.

***Polymyositis:** NO cutaneous involvement

Dermatomyositis: BOTH muscular and cutaneous abnormalities.

Diagnostic approach:

1- EMG
2- MRI: displays inflamed muscles
3- Muscle biopsy: most reliable diagnostic test showing varying stages of necrosis & regeneration with inflammatory infiltrate.
4-Skin biopsy is suggestive but not diagnostic that shows interface dermatitis.
Polymyositis: CD8+, **DM** : CD4+

Dusky red rash in face and elbow

Gottron sign

Labs:

Sed rate & CRP elevated in 50%
Creatine kinase (CK) is the best marker
24 hour urine creatine level >200 mg is diagnostic
Other enzymes : AST,LDH

Serology:

Myositis-specific antibodies (MSA) : in <50% of patients.
Anti Jo-1 (antisynthetase): in 20%, with lung disease & Raynaud phenomenon.

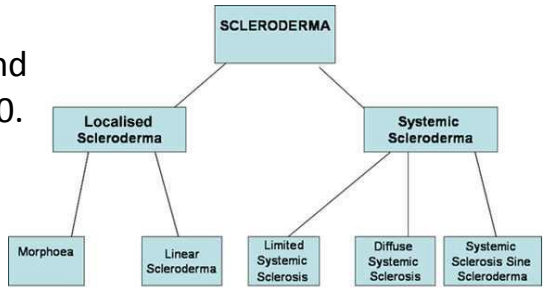
Treatment:

Corticosteroids : PO and taper, or pulse therapy, monitoring CK.
Immunosuppressants: M

ethotrexate,azathioprine.
Cyclophosphamide & cyclosporine are less effective
IVIG: resistant cases, children with vasculitic component &steroid induced DM
Antimalarials and topical steriods : helps cutaneous lesions, topical steroids, sunscreen.
Paraneoplastic: appropriate Rx of underlying malignancy.
Physiotherapy to improve strength and flexibility of the muscles

Scleroderma (Skin Sclerosis):

A group of rare diseases that involve the hardening and tightening of the skin and connective tissues
. Scleroderma affects women more often than men and most commonly occurs between the ages of 30 and 50.

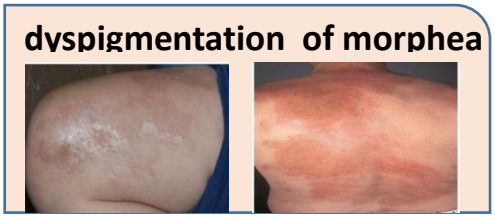


Morphea :

Sclerosis w/o systemic involvement . Uncommon.F>M 3:1, incidence around
2/100000 per year. Poorly understood, association with *Borrelia Burgdorferi* , circulating autoantibodies support immune role.

Classic morphea:

-Circumscribed sclerotic plaque with ivory center & red violet periphery (lilac ring).*Sometimes in linear Distribution on face and extremities.
-It subsides on its own over time leaving dyspigmentation and scars.Cutaneous -Systemic findings uncommon, raely malaise or Raynaud.



Variants:

Plaque form. Atrophoderma of Pasini & Pierini
Guttate ,Nodular
Linear. (Coup de sabre)
Hemifacial atrophy (Parry-Romberg syndrome)
Lichen sclerosis et atrophicus- like lesions

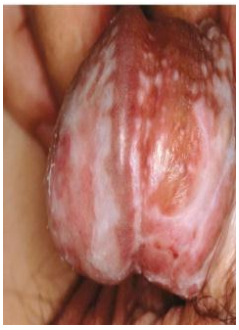
Diagnostic:

Skin biopsy can confirm diagnosis.
ANA and anti ssDNA may be found in linear widespread morhea

Therapy:

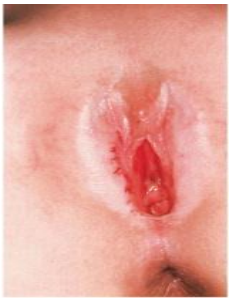
High potency topical corticosteroids.
PUVA or UVA

Lichen sclerosis :



Dermatosis that present with either porcelain-white papules & plaques or atrophy.
F>M

-similar to morphea, smaller white papules (en confetti) on trunk, especially upper back



-Genitals: ***women***: white atrophic lesions over vulva, labia minora,clitoris may spread to perianal, sometimes mistaken for abuse, pruritic ++.

Men: common cause of phimosis, usually NOT pruritic, resolves after circumcision

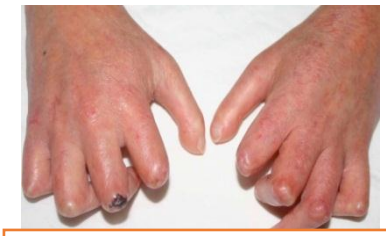
N.B: slight risk for Small cell carcinoma, to be monitored.

Therapy:

High potency topical corticosteroids

Topical immunomodulators (tacrolimus or pimecrolimus) for long term maintenance.

Systemic Sclerosis:

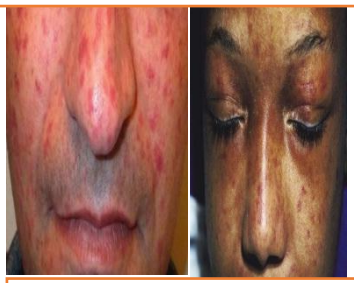


***puffy hands, sclerodactyly, tightening, reduced motion,** fingertip ulcers, calcinosis.

multiorgan disease with diffuse sclerosis of connective tissue favoring skin, lung, GI & kidneys. F>M 5:1, but men have worse Prognosis .Incidence: 1-2/100000 per year .

Pathogenesis:

Genetic predisposition (HLA-DR3,-DR5..).
Vessel damage as primary event.Increased production: type I,III & IV Collagen, fibronectin & Proteoglycansin affected connective tissue.



***microstomia, reduced facial expressions.**

Classification :

Acral sclerosis types I,II (limited)
Systemic sclerosis: starts on trunk, severe facial involvement.

CREST syndrome:

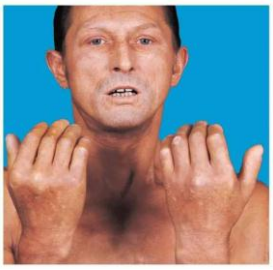
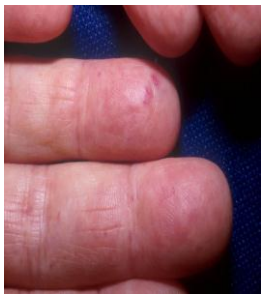
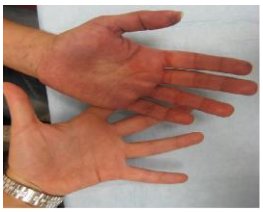
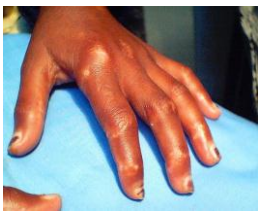
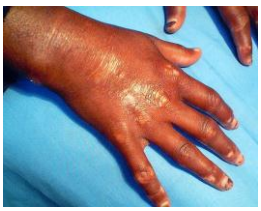
Calcinosis, ***R***aynaud phenomenon, ***E***sophageal disease, ***S***clerodactyly, Telengiectasia.

Clinical features:

*Hands*Face**
Diffuse sclerosis: sometimes restriction of respiratory motion.
Raynaud phenomenon:Confetti like hypopgmentation: mistaken for vitiligo, more dramatic in blacks.

Systemic Sclerosis

Systemic findings:
GI tract: reduction tongue motility, swallowing problems, sicca syndrome, impaired esophageal motility.
Lungs: skin restricts motion, pulmonary fibrosis.
Kidneys: Hypertention, nephrosclerosis.
Heart: subtle & late; MC fibrosis, pericardial effusion.
Liver: primary biliary cirrhosis.
Musculoskeletal: arthralgias, muscle atrophy
Bones: acro-osteolysis
Teeth: widening of periodontal membrane

Diagnostic approach:
Diagnosis is made based on clinical features and presentation.
Skin: biopsy will show skin atrophy with preservation of skin appendages.
Lungs: CXR, pulm diffusion studies
Skeleton: X.Ray for osteolysis, calcifications.
GI: esophageal manometry, scintigraphy
Heart: ECG,echo
Kidneys: renal function, urine status

Labs:
Routine (CBC, LFT, sed rate, CRP)
Serology:
ANA (>90%).
Anti topoisomerase I (Anti SCL-70)(30-70%)
Anti RNA (<5%); poor outlook
Anti-centromere ab : in type I & CREST.
Anti-DM-Scl in overlap
Rheumatoid factor (+ in 30%)

Therapy:

Raynaud: NO smoking,avoidance of cold, Ca channel blocker (nifedipine,diltiazem..)
Calcification: surgery
Ulcers: occlusive dressing
Sclerosis: systemic Corticsteriods, perhaps combined with azathioprine. MTX, cyclosporine, cyclophosphamide can be tried.
Aspirin: pain relief and inhibition of platelet function
Calcinosis cutis:Nifedipine, surgical or laser excision.
Skin sclerosis or contracture: physiotherapy, phototherapy.
GI: proton pump inhibitor, surgery for strictures.
Kidney: ACE inhibitors.
Lung: Prostacycline for pulmonary HT, cyclophosphamide for interstitial lung disease.
In severe cases: immunosuppressant, D-Penicillamine might be used.

Important table from team 432*

Very Important

Table 2: Useful Antibodies for CTD-ILD Assessment

Autoantibody	Associated CTD
High titer ANA (>1:320 titer)	Many
High titer RF (>60 IU/mL)	RA, Sjögren's disease, SLE
Anti-CCP	RA
Anti-centromere	Systemic sclerosis
Anti-nucleolar-ANA	Systemic sclerosis
Anti-Ro (SS-A)	Many
Anti-La (SS-B)	SLE, Sjögren's disease
Anti-Smith	SLE
Anti-ribonucleoprotein	SLE, MCTD
Anti-dsDNA	SLE
Anti-topoisomerase (Scl-70)	Systemic sclerosis
Anti-tRNA synthetase antibodies	Poly-/dermatomyositis (antisynthetase syndrome)
Anti-PM-Scl	Systemic slerosis/myositis overlap
Anti-Th/To	Systemic sclerosis
Anti-U3 ribonucleoprotein	Systemic sclerosis
ANCA panel	Systemic vasculitis

Done By:

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